

Oro-facial presentations of CKD The Forgotten Story

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Types of facial skeletal changes

1. Osteitis fibrosa cystica & brown tumours (commonest)
2. Fibrous dysplasia like lesion
3. Leontiasis ossea (rarest)



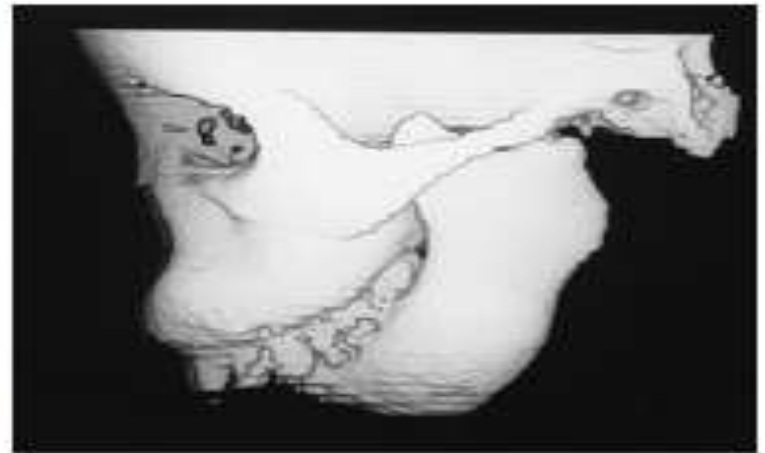
In 1991, a 16-year-old white man with dialysis-dependent renal failure since age four years (from sclerosing glomerulonephritis) first developed signs of maxillary overgrowth. He suffered other clinical complications of rickets and renal osteodystrophy including bilateral slipped capital femoral epiphyses and extensive soft tissue calcifications and was maintained on continuous ambulatory peritoneal dialysis. By age 18 years, facial changes had progressed dramatically (A, age 12 years, B and C, age 18 years). Intact parathyroid hormone levels ranged from 245–3165 pg/ml (normal range 13–64 pg/ml), and alkaline phosphatase levels from 698–1744 U/L (normal range 20–96 U/L). CT and MR imaging confirmed marked hypertrophy and hyperostosis of the maxilla and mandible with diffuse, heterogeneous widening of the diploic space (D, three-dimensional recon-



A



B



D



R

L

A 46-year-old woman with known epilepsy, on long-term phenobarbitone therapy, developed end-stage renal failure. She had been on maintenance hemodialysis for 2 years when she became noncompliant with oral α -calcidiol therapy. She subsequently developed bulging of the malar bones and widening of the nares and interdental space (Figure 1). Her intact parathyroid hormone was 230 pmol per liter, and a skeletal survey was consistent with renal osteodystrophy. A radiograph (Figure 2) and a computed tomographic scan of the skull (Figure 3) showed

hyperostosis of facial and cranial bones (Figure 2, arrows) and ground-glass expansion of the maxilla, mandible, and skull vault with a 7×2.5 cm brown tumor arising from the maxilla (Figure 3, arrows). These findings are characteristic of uremic leontiasis ossea. Parenteral vitamin D was commenced for the patient, and she is awaiting parathyroidectomy. It is important

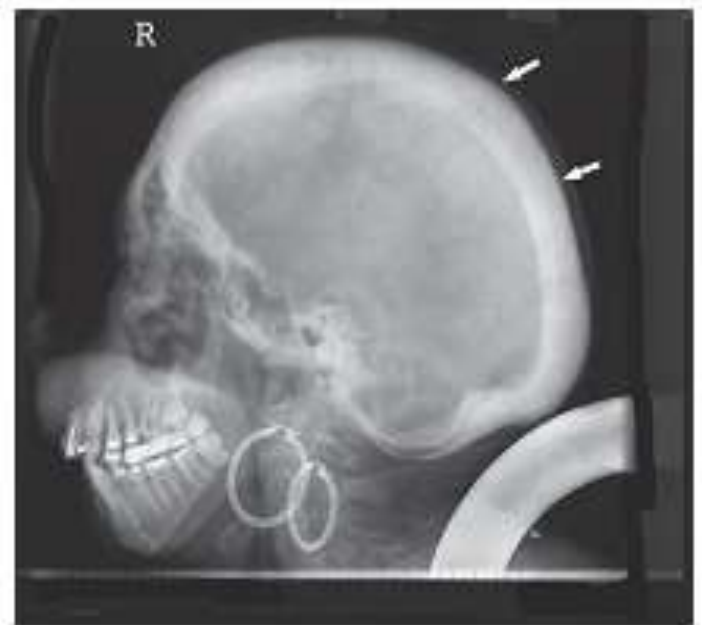


Figure 2 | Plain radiograph of the skull, revealing hyperostosis of the facial and calvarial bones (arrows).



Figure 3 | Computed tomograph of the skull, revealing brown tumor of the maxilla (arrows).

Leontiasis Ossea

It is important to recognize features of uremic leontiasis ossea, as it may result in:

- Life-threatening upper airway obstruction
- Compressive cranial neuropathy.

Saglikler Syndrome (The Big Failure)

- Introduced in 2004 in patients with ESKD and severe SHPT
- This syndrome describes maxillary and mandibular deformities, dental abnormalities, benign soft tissue tumors in mouth, and various kinds of skeletal changes including short stature and fingertip abnormalities.



Symptoms Reported to Coexist With Musculoskeletal Manifestations of Sagliker Syndrome

Study	Number of Patients	Symptom	Prevalence
Erkan et al ¹⁹	10	Hearing loss	60%
Ozenli et al ²⁰	13	Depression disorder	46%
Ozenli et al ²⁰	13	Anxiety disorder	23%

Is survival enough for quality of life in Sagliker Syndrome-uglifying human face appearances in chronic kidney disease?

Conclusion: SS is a serious and severe complication of CKD. Late and improper treatment leads to abnormalities throughout skeleton particularly in the skull and face. Changes particularly in children and teens become irreversible-disastrous for appearance and psychological health. Appropriate treatment must begin as early as possible in specialized centers. It is possible that SS patients may survive long-term with dialysis, but with all those particular changes could anyone claim this type of life would continue in an acceptable way without extending their height, correcting all the changes in the skull and face, remodeling new faces and most particularly convincing the patients to deal with all those tragedy-dramatic psychological problems?

Fibrous Dysplasia

Case Report

A 37-year-old man with a history of end stage renal disease who was on hemodialysis for 5 years presented with an expanding palatal lesion of 6-months' duration, which caused dysarthria and oral dysphagia. He denied any other regional symptoms. Physical examination revealed a firm enlarged nontender hard palate that had descended to the occlusal plane. Combined with extensive maxillary hypertrophy bilaterally, the palatal changes caused the maxillary teeth to splay outwards (Fig 1).



Localized Jaw Enlargement due to Fibrous Dysplasia in a Hemodialysis Patient

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Figure 2: CT of maxilla and cranium shows a mass of ossified filling and distorting the left maxillary sinus measuring 5 x 4 cm



Case Description: A 24-year old, female, Caucasian patient presented chronic glomerulonephritis recurrence and lost the transplanted kidney five years before, undergoing arteriovenous fistula hemodialysis three times a week. She presented swelling of the left masseter area with a hard consistency on palpation, covered by intact skin, swelling at the bottom of the left atrium, with a hard consistency on palpation, a mucosa-like color and absence of inflammation signs, suggesting expansive bone lesions on the face. These features were compatible with hyperparathyroidism brown tumor and/or osteodystrophy. The CT scan showed expansive bone lesions of heterogeneous appearance on the left jaw, maxilla/nasal floor, and right frontotemporal suture areas. The clinical and histopathological characteristics of the lesion, in association with PHT hormone high serum levels led to renal osteodystrophy diagnosis. The patient was referred to the nephrology services.

OFC(brown tumour)



Fig. 1: Three-dimensional reconstruction in CT scan: an expansive area in the left mandible extending from the alveolar ridge to the base of the mandible in the molar region was observed.

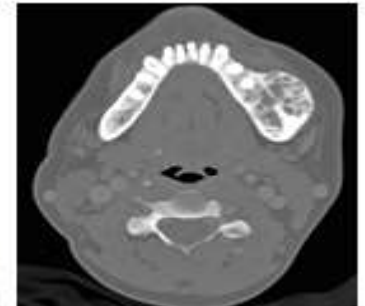


Fig. 2: CT axial cut with bone window: expansive area of heterogeneous density in the left mandible.



Fig. 3: Three-dimensional reconstruction in CT scan: an expansive area in the anterior mandible, nasal floor, left side of the jaw and frontoparietal suture was observed.

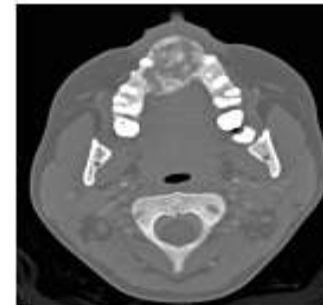


Fig. 4: CT axial cut with bone window: expansive area of heterogeneous density in the anterior mandible.



Fig. 5: Three-dimensional reconstruction in CT scan: expansive area in the right frontoparietal suture.

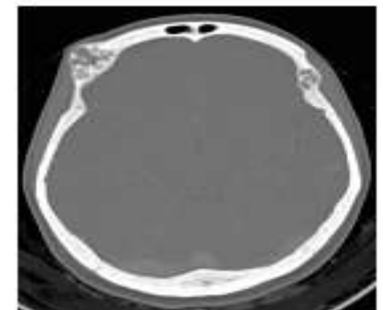


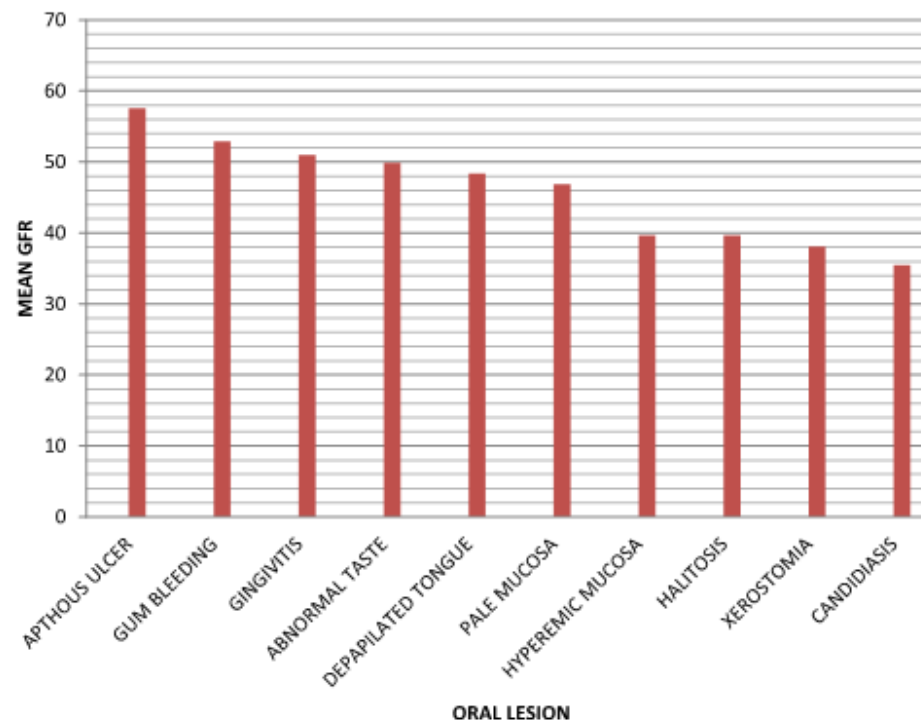
Fig. 6: CT axial scan: areas of heterogeneous bone expansion in the right and left frontoparietal suture.

RESEARCH ARTICLE

Open Access

Oral findings in chronic kidney disease: implications for management in developing countries

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Drugs

1. **Bisphosphonates**-----> Avascular Jaw necrosis

- **Criteria to diagnose:**

1. The patient possesses an area of exposed bone in the jaw persisting for more than 8 weeks,
2. The patient must present with no history of radiation therapy to the head and neck,
3. The patient must be taking or have taken bisphosphonate medication.

Contd.,

2. CNI, nifedipine -----> Gingival hyperplasia(>3m ttt & children)

3. Anticoagulants-----> Gingival bleeding

4. Diuretics, BBs-----> lichenoid lesions

International evaluation of unrecognizably uglifying human faces in late and severe secondary hyperparathyroidism in chronic kidney disease. Sagliker syndrome. A unique catastrophic entity, cytogenetic studies for chromosomal abnormalities, calcium-sensing receptor gene and GNAS1 mutations. Striking and promising missense mutations on the GNAS1 gene exons 1, 4, 10.

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Gene Mutations in Chronic Kidney Disease Patients With Secondary Hyperparathyroidism and Sagliker Syndrome

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THANK YOU

على قدر الهدف يكون الانطلاق

ففي

" طلب الرزق قال: " فامشوا

" وللصلاة قال: " فاسعوا

" وللجنة قال: " وسارعوا

" وأما إليه فقال: " ففروا إلى الله